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Prion Diseases: Adapting and Collaborating

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Transmissible spongiform encephalopathies or 'prion diseases' include Creutzfeldt Jakob disease (CJD) in humans and various conditions in animals. These emerging diseases pose a challenge to health care and public health staff for many reasons including:

- very long incubation periods (on the order of years to decades)
- no treatment and invariably fatal outcome
- no definitive diagnosis clinically or with routine lab tests so autopsy is needed to confirm the diagnosis
- no known cause for most cases
- resistance to disinfection so infection control is needed during invasive neurological procedures
- remaining gaps in understanding

Collaboration for Diagnosis

To pool public health experience and skills, the Washington State Department of Health (DOH) has partnered with neighboring state health agencies (Idaho and Alaska), the Centers for Disease Control and Prevention, and the National Prion Disease Surveillance Pathology Center (NPDSPC) with its Cleveland-based reference laboratory. This collaboration will enhance the recognition and laboratory confirmation of CJD, the major human prion disease.

The NPDSPC runs a unique public health program to minimize barriers to confirmatory laboratory diagnosis of prion diseases. With a family's consent, NPDSPC provides brain autopsy including transport of the body so a pathology center can remove brain tissue, transport of the tissue to the reference laboratory, and advanced molecular and immunohistochemical laboratory testing – all free of charge.

In Washington State, CJD and other prion diseases of humans fall under the notifiable condition category 'Rare diseases of public health significance.' Washington's CJD rate is consistent with that of the United States at 1-2 cases/million population annually so the disease is expected to affect only a few Washington residents each year.

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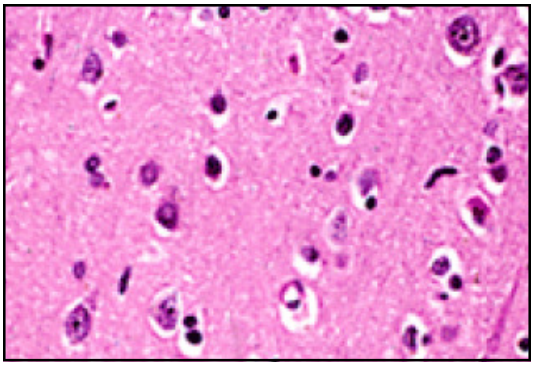
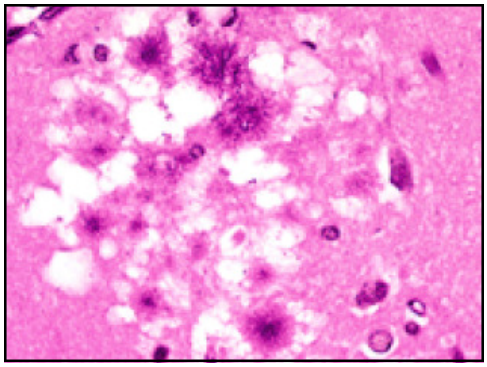
Animal prion diseases such as bovine spongioform encephalopathy of cattle ("mad cow disease"), scrapie of sheep and chronic wasting disease of elk and deer are managed by Washington State's Department of Agriculture and the Department of Fish and Wildlife.



Normal human cerebral cortex showing no significant pathological changes



nvCJD cerebral cortex showing the "florid" plaques that consist of vacuoules (spongiform degeneration) containing amyloid plaques

Images courtesy of InPro Biotechnology, Inc., and Stephen J. DeArmond, M.D., Ph. D., Professor of Pathology (Neuropathology), Department of Pathology, University of California San Francisco.

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http://listserv.wa.gov/archives/epitrends.html

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New Public Health Activities

DOH has a new website that highlights the surveillance and reporting guidelines for notifiable human prion diseases: http://www.doh.wa.gov/notify/nc/prion.htm

The site includes a fact sheet for health care providers and a case report form. A document answering frequently asked questions will be available soon.

In December 2007, DOH sent a letter to Washington State licensed neurologists, pathologists, and health officers asking for assistance in reporting suspected CJD cases and promoting autopsy for laboratory confirmation of cases.

The CJD Foundation is a resource to support the families of affected individuals: http://www.cjdfoundation.org/index.html . The Foundation also has a new 55 minute DVD "Confronting CJD and Other Prion Disorders" available for continuing medical education (CME): http://www.cjdfoundation.org/medical.html

Human prion diseases challenge the usual communicable disease surveillance system. However these diseases merit attention and together public health agencies can provide the resources and skills to track these conditions.

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Annual Communicable Disease Report, 2006

The 2006 annual Communicable Disease Report is now available online: http://www.doh.wa.gov/notify/list.htm

In addition, a limited number of copies will be printed for libraries, local health jurisdictions, and other state offices. Copies will be mailed at the beginning of the year. Highlights of the annual report follow.

Foodborne Illness

Although most reported cases of foodborne infections are sporadic, outbreaks require considerable involvement of public health agencies. During 2006 several local health jurisdictions collaborated on multi-state outbreaks, one involving *Salmonella* Tennessee linked to contaminated peanut butter and one involving *E. coli* O157:H7 associated with spinach.

In 2006 a large outbreak of vibriosis (*Vibrio parahaemolyticus* infection) was associated with raw oysters harvested in Pacific Northwest waters. The vibriosis rate was the highest in Washington in the past two decades.

Among other enteric and foodborne outbreaks investigated by local health jurisdictions were an outbreak of *E. coli* O157:H7 infections related to raw milk from a commercial dairy and three scombroid toxin outbreaks related to contaminated fish. A single case of trichinosis was reported associated with consumption of raw cougar meat.

West Nile Virus

West Nile virus was first identified in the United States in 1999 and subsequently spread across the continent. Although birds and horses with West Nile virus infection and Washington residents infected during out-of-state travel were detected in Washington as early as 2002, the first endemically acquired cases of human West Nile virus (WNV) infection were detected in 2006.

Sexually Transmitted Infections

As in previous years, sexually transmitted infections are the most commonly reported notifiable conditions in Washington State. There were 17,819 *Chlamydia trachomatis* infections reported in 2006 (279.5 cases/100,000 population) which continued an increase in rates since 1996. Compared to previous years the rates of primary and secondary syphilis (2.9 cases/100,000) and of gonorrhea (66.4 cases/100,000) also continued to rise.

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Pertussis

In 2006 the rate of pertussis (5.9/100,000) reached the lowest point since 2001, reversing a trend of highly elevated rates during recent years. The 2006 rate was lower than the last reported national rate, 8.7/100,000 in 2005. Although 60% of pertussis in Washington occurs among those 10 years of age and older, the highest rate and the most serious illnesses continue to occur among children under one year of age.

Mumps

During 2006, using more stringent mumps reporting guidelines instituted after an outbreak in the Midwest, seven confirmed and 34 probable mumps cases were identified in Washington. No cases were linked to the national outbreak. Eleven case patients had traveled during their likely exposure period, including two to Europe, one to the Bahamas, one to British Columbia, and one to the Philippines.

Cryptococcus gattii

An unusual agent, *Cryptococcus gattii*, may cause respiratory illness or meningitis. Unlike the closely related opportunistic species *C. neoformans, C. gattii* primarily affects immunocompetent persons, although smoking or treatment with systemic steroids may increase a person's risk. Infection occurs by breathing spores of the fungus, which has been identified from environmental sources such as trees and soil. Vancouver Island in British Columbia, Canada found both human and animal cases of *C. gattii* infection beginning in 1999. During 2005, three cats living in Washington near the border but without exposures in Canada were diagnosed with *C. gattii.* Two Washington residents with *C. gattii* disease in 2006 may have been locally exposed but did have travel out of state as well.

Notifiable Conditions Surveillance

Notifiable conditions surveillance in Washington State depends on reporting of cases to local health jurisdictions so appropriate public health interventions can be done. Disease surveillance has improved with newer electronic systems but still relies on health care providers, health care facilities, laboratories, veterinarians, and other reporters contacting public health agencies when cases are identified.

The Department of Health is grateful to all the disease investigators and reporters whose work during the past year has contributed to this annual summary.